CASE REPORT

Congestive hepatopathy secondary to large renal arteriovenous malformation

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SUMMARY

A 75-year-old woman presented with acute onset dyspnoea, and was found to have signs of pulmonary congestion on clinical examination. Imaging revealed cardiomegaly and coincident congestive hepatopathy, secondary to a left renal arteriovenous malformation. The presence of a high flow vascular shunt in the left kidney was possibly the causative factor behind both the high-output cardiac failure and congestive hepatopathy.

BACKGROUND

This report presents a case of congestive hepatopathy of a non-cardiogenic aetiology. This is a rare entity since congestive hepatopathy, or cardiac cirrhosis, is commonly known to arise in the setting of chronic heart disease. In this case, a large left renal arteriovenous malformation (AVM) was found to be the cause of the liver congestion, which also brings to light an unusual complication of AVM. The appearance of the above findings on CT scan was described. Moreover, the pathogenesis and management of cardiac cirrhosis in this case are discussed.

CASE PRESENTATION

A 75-year-old woman presented to the hospital with new-onset severe dyspnoea. Patient denied previous history of progressive dyspnoea on exertion or exercise intolerance prior to presentation, but she did report generalised fatigue; however, it did not interfere with her every day functioning. The patient was a non-smoker, her medical history was negative for prior hospital admissions. The patient did not report any nausea, abdominal pain nor flank pain. Her blood pressure readings were within normal, but she was mildly tachycardic. On examination, she was found to have signs and symptoms of pulmonary congestion. No other clinical findings were of significance; no extra heart sounds were heard, no hepatomegaly and no oedema of lower extremities.

INVESTIGATIONS

Alanine aminotransferase (ALT), aspartate aminotransferase (AST), alkaline phosphatase, bilirubin and albumin levels were taken and were found to be within the normal range.

Her work-up included an abdominal CT scan with intravenous contrast. Images revealed an irregular contour of the liver. The arterial phase showed a patchy pattern of enhancement which became homogenous on the delayed phases (figure 1); these findings are typical of cirrhosis.

Dilated tubular and tortuous vessels with arterial enhancement were identified in the left kidney.

They involved the left renal hilum and were continuous with the single left renal artery and left renal vein. The latter was significantly dilated and was found to be impinging on the abdominal aorta (figures 1–4); its insertion onto the inferior vena cava (IVC) was seen in the proximity of the branching of the hepatic veins. The IVC was also significantly dilated presumably due to the increased blood flow from the left renal vein which showed enhancement similar to that of the abdominal aorta (figure 1). These findings are consistent with abnormal left renal vascular malformations or arteriovenous fistulas. In addition, some of the tortuous vessels within the left kidney exhibited focal aneurysmal dilation with rim calcifications (figures 2 and 4). The left renal cortex was preserved.

On the scout image, the cardiac silhouette was significantly enlarged, which is secondary to high output cardiac failure. This was confirmed by an echocardiogram.

DIFFERENTIAL DIAGNOSIS

This case involves an elderly patient who presented with a picture of cardiac failure, and was found to have cirrhotic liver changes on imaging. Combined cardiac and liver abnormalities can occur in several settings including: heart disease causing the liver abnormality, liver disease affecting the heart, or both disorders having a common aetiology.¹

Congestive heart failure commonly impacts the liver, it can result in elevation of liver enzymes which can be significant and rapid in the setting of cardiogenic ischaemic hepatitis;¹ however, our patient has normal levels of liver enzymes. Furthermore, the clinical presentation and the description of liver changes here are indeed suggestive of congestive or cardiac cirrhosis; but the fact that both abnormalities presented to us simultaneously makes this unlikely and suggests a joint cause. Cardiac cirrhosis usually constitutes a hepatic reaction to chronic increased venous pressure and hypoxia caused by long-standing heart problems such as ischaemic disease or cardiomyopathies.^{2,3}

As a matter of fact, imaging did reveal a large left renal vascular malformation with its main outflow vessel located close to the hepatic veins. Renal vascular malformations have been reported to cause high-output heart failure;⁴ and in the scenario of this patient, it might have resulted in the development of congestive hepatopathy as well.

OUTCOME AND FOLLOW-UP

The patient was offered the option of endovascular occlusion of the fistula however she declined it. She

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Figure 1 (A and B) Coronal and curved reformat images illustrate the relationship of the enlarged left renal vein (black arrow) and the proximity of its insertion to the infrahepatic portion of the dilated inferior vena cava (white arrowheads). The density within the left renal vein approaches that of the aorta (black star), which is consistent with arteriovenous shunting. Also noted is the continuous communication with the dilated tortuous vessels occupying the left renal pelvis (white arrow). The liver contour (black arrowheads) appears lobulated in (B) with slight prominence of the caudate lobe (white star) which are changes typical of cirrhosis.

Figure 2 Axial contrast-enhanced CT scan image obtained at the level of the left renal vein (black arrow) reveals its dilatation. The tortuous vessels in the left kidney exhibit focal aneurysmal dilatation and rim calcifications (star). The white arrowheads point at the dilated inferior vena cava.

Figure 3 Sagittal reformat CT image illustrates the enlarged left renal vein as it courses anterior to the aorta, causing an impression on it (black arrow).

was started on medical treatment for her cardiac condition which included an ACE inhibitor and a diuretic; patient symptoms improved shortly afterwards and she was advised to commit to regular close follow-up. However, the patient was lost to follow-up after a while, and her current clinical status is unknown.

DISCUSSION

Cardiac cirrhosis is a commonly recognised entity in which patients present with imaging features of liver cirrhosis in the setting of chronic heart disease. Although cardiac cirrhosis is the term most commonly used, congestive hepatopathy and chronic passive liver congestion are more accurate ways of referring to the disease.⁵

Cardiac cirrhosis, or congestive hepatopathy, involves a range of morphological changes that represent the response of liver parenchyma to the interplay of several factors including increased venous pressure, hypoxia and thrombosis. The main causes of cardiac cirrhosis that have been commonly reported in the literature include: ischaemic heart disease (31%), cardiomyopathy (23%), valvular heart disease (23%), restrictive lung disease (15%) and pericardial disease (8%).² In the setting of chronic heart disease, the impairment in cardiac output and clearance lead to increased IVC pressure, increased venous pressure and subsequent venous congestion. This gets transmitted via the hepatic veins to the liver sinusoids resulting in their dilatation and engorgement with blood; consequently, the liver becomes enlarged and swollen. Additionally, deoxygenated blood continues to accumulate in sinusoids secondary to the poor circulation, causing relative ischaemia. Sinusoidal stasis may also lead to thrombosis of the sinusoids and terminal hepatic venules, which if propagates to medium-sized hepatic veins and portal vein branches can lead to ischaemia.^{1 6}

Therefore, the chronic stasis or passive congestion will result in centrilobular hepatocyte injury, necrosis, collagen deposition and eventually fibrosis with the typical cirrhotic appearance of the liver.^{6 7}

In this case, the patient presented with a compromised cardiac condition concomitant with changes of liver cirrhosis that were found to be secondary to the presence of a large left renal vascular malformation whose main outflow vessel was located very close to the hepatic circulation, thereby possibly precipitating both conditions. Although, AVMs are known to cause high-output heart failure, especially those large in size,⁴ this particular

Figure 4 (A–C): All are three-dimensional MIP reformatted images that clearly show the dilated left renal artery (black arrow) and its relationship to the dilated tortuous vessels occupying the left renal pelvis (white arrow). The outline of the left kidney (white arrowheads) can be differentiated from the vessels in image (C). The large structure protruding from the lower aspect of the renal pelvis (star) is consistent with focal aneurysmal dilation of the shunted vessels.

occurrence was not found to be reported in the literature, after searching the Medline or Embase databases. However, Naschitz *et al*¹ does touch on joint causes of hepatic and cardiac dysfunction, this specific aetiology was not mentioned.

Renal vascular malformations are rare; studies involving patients undergoing evaluation with vascular imaging techniques estimate the rate to be as low as one case per 1000–2500 patients.⁸ These could be classified into either congenital AVMs or acquired arteriovenous fistulas (AVF) which are commonly iatrogenic or post-traumatic. These two types can be differentiated according to their appearance on imaging.⁹

AVMs, despite being congenital anomalies, tend to manifest after the fourth decade of life. In addition, they have a distinct predilection to female patients.¹⁰ This picture is reinforced by the scenario we have here. Furthermore, reports differ concerning the side that is most commonly involved, or whether one versus multiple lesions are usually present.¹⁰

Congenital vascular malformations are further divided into three types: cirroid, angiomatous and aneurysmal. Cirroid type is the most commonly recognised form; it is characterised by the presence of a vascular nidus with multiple feeding arteries and draining veins, and is located near the collecting system. The angiomatous type has a single large feeding artery with multiple distal branches and draining veins, and it is usually peripheral or cortical.^{9–11} Third is the aneurysmal type which is also known as idiopathic AVM. It is composed of a single draining artery and a single draining vein with aneurysmal dilation.

AVF are more common and lack a vascular nidus. Acquired fistulas can be ruled out by excluding previous penetrating trauma, percutaneous biopsy, surgery such as nephrectomy, malignancy or inflammation in the clinical history of the patient.^{9–12}

Aneurysmal AVMs can be difficult to differentiate from AVFs; in fact, congenital AVFs are considered in some references as

part of the spectrum of congenital AVMs. It is of relevance to note that both aneurysmal AVMs and congenital AVFs are high-flow shunts with enlargement of the affected vessels.

The patient in this case has no history of prior trauma or intervention involving her left kidney which excludes iatrogenic AVF. Keeping in mind the single vascular origin and drainage of the left renal vascular mass and the lack of a vascular nidus, it can be either a congenital AVF or aneurysmal AVM; both forming a high flow shunt. The involvement of the left renal vein by the high flow shunt as well as its proximity to the hepatic veins as it inserts into the IVC have possibly reproduced the conditions for the development of congestive hepatopathy; referring here to the increased IVC pressure also seen in cardiac failure. It is likely that the left renal AVF or aneurysmal AVM is the principal causative factor of both liver cirrhosis and high output heart failure.

Patients with renal AVM usually present clinically with flank pain, haematuria and hypertension or as in this case high output cardiac failure and pulmonary congestion.¹²

Patients with congestive hepatopathy usually do not present with any additional manifestations apart from signs and symptoms of right heart failure attributed to their primary cardiac condition.³ In our patient, as in the majority of patients, ALT, AST, alkaline phosphatase, bilirubin and albumin levels were within the normal range. Treatment of cardiac cirrhosis classically targets the heart failure. In other terms, it involves dealing with the cause of elevated right-sided heart pressure and hepatic venous congestion referring here to the left renal AVM.⁷

Management of vascular malformations can be individualised since expectant measures with mainly observation are allowed in some cases, but risk of later complications is not nil here. On the other hand, patients presenting with heart failure, uncontrolled hypertension, intraparenchymal haemorrhage or other threatening manifestations require definitive treatment.⁴ The first choice consists of angiographic embolotherapy using either alcohol ablation in cases with multiple feeding vessels and a nidus,¹³ or coil embolisation for the larger AVFs.¹² The latter was the option offered to our patient; however she declined it. In order for any intervention to be successful and to minimise chances of recurrence, it should be paralleled with medical management of the heart failure, hypertension and haemorrhage if present.⁸

While nephrectomy can cure AVMs, it is reserved for cases refractory to embolotherapy or those associated with malignancy; this is so mostly due to the fact that a significant amount of healthy renal tissue is removed in surgery. Moreover, surgery can typically result in a wide range of complications, such as cardiac arrhythmias from electrolyte disturbances, surgical bleeding or hypotension resulting in ischaemic complications.⁸

In addition, there are the well-known consequences of several surgical procedures, including pulmonary, gastrointestinal and infectious consequences.

Learning points

- ▶ Congestive hepatopathy can be the result of non-cardiogenic aetiology.
- ▶ Hepatic and cardiac disorders can arise from common aetiology.
- ▶ Congestive hepatopathy is a possible complication of renal arteriovenous malformations (AVMs).
- ▶ Treatment should address the AVM to target both conditions.

Competing interests None.

Patient consent Obtained.

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